Role of 3D power Doppler sonography in early prenatal diagnosis of Galen vein aneurysm

3D power Doppler sonografinin Galen ven anevrizmasının erken prenatal tanısındaki rolü

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Abstract

Vein of Galen aneurysm malformation (VGAM) is a rare congenital vascular anomaly. Although the cause of VGAM remains to be elucidated, the current hypothesis is persistence of the embryonic vascular supply, which leads to progressive enlargement and formation of the aneurysmal component of a typical VGAM. Here, we present a 36-year-old woman at 23 weeks' gestation (gravida 3, para 2) who was evaluated using 3D power Doppler sonography for the prenatal diagnosis of a vein of Galen aneurysm. Investigation using 3D power Doppler sonography allowed for a non-invasive yet diffuse and detailed prenatal assessment of VGAM. Thus, we suggest that prenatal sonography with 3D power Doppler may be an option in cases of VGAM. (J Turkish-German Gynecol Assoc 2013; 14: 178-81)

Key words: Galen vein aneurysm, power Doppler, 3D sonography

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Özet

Galen ven anevrizma malformasyonu (VGAM) nadir görülen bir konjenital vasküler anomalidir. VGAM nedeni tam olarak aydınlatılmış olmamasına rağmen, güncel hipotez, tipik anevrizmanın progresif genişlemesine yol açan embriyonik vasküler desteğin persiste etmesidir. Bu çalışmamızda 36 yaşında ve 3D power Doppler sonografi kullanarak Galen ven anevrizması değerlendirilen 23 haftalık gebeliği (gravida 2, para 2) bulunan bir olguyu sunduk. VGAM'nin prenatal değerlendirmesinde non-invaziv yoldan geniş ve ayrıntılı bilgi sağlamak için 3D power Doppler sonografi kullanımı uygun görülmektedir. Bu nedenle VGAM bulunan vakalarda 3D power Doppler sonografiyi opsiyonel bir seçenek olarak önermekteyiz. (J Turkish-German Gynecol Assoc 2013; 14: 178-81)

Anahtar kelimeler: Galen ven anevrizması, power Doppler, 3D sonografi

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Introduction

Vein of Galen aneurysm malformation (VGAM) is a rare congenital vascular anomaly that generally manifests in the neonatal period with symptoms of congestive heart failure. Hydrocephalus-related obstructive effects, seizures, encephalomalacia, and subcortical calcification may be present during the infantile period (1-4). In paediatric and adult patients, VGAM is rare and presents with headache and symptoms of subarachnoid haemorrhage (5, 6). VGAM has been reported to comprise 30% of paediatric vascular malformations (7).

Although the cause of VGAM remains to be elucidated, the current hypothesis regarding the possible mechanism is persistence of the embryonic vascular supply known as the choroidal arteries and the anterior segment of the median prosencephalic vein of Markowski, which normally regresses. This persistence leads to progressive enlargement and formation of the aneurysmal component of a typical VGAM (8).

Currently, the widespread utilisation of foetal sonography has resulted in the frequent diagnosis of this rare vascular malformation. The most common diagnostic criterion on real-time imaging is the detection of an intracranial cystic structure. Doppler sonography with colour flow imaging makes it easier to identify the blood flow within the cyst. Recent investigations of intracranial vascular lesions by using the 3D power Doppler mode reveals some benefits compared to conventional colour Doppler sonography, such as obviating aliasing distortions, angle dependence, overwhelming noise, and more detailed anatomical correlation. Cranial examination may also reveal mild to moderate hydrocephalus at advanced gestational ages. Jugular vascular ectasia and cardiomegaly with congestive heart failure and its peripheral findings, such as ascites, may be seen in the affected foetus or neonate due to high-velocity flow in this arteriovenous (AV) fistula and are accepted as the main factors determining prognosis (9).

In this case report, we present the prenatal diagnosis of a VGAM and evaluate it using the 3D power Doppler mode.

Case Report

A 36-year-old woman at 23 weeks' gestation (gravida 3, para 2) was referred to our prenatal diagnosis unit for further

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evaluation of corpus callosum agenesis. Her obstetric and medical history was unremarkable except for the presence of Rh incompatibility. Indirect Coombs test was negative in the second trimester evaluation. The triple test during the 16th week revealed a 1/81 risk for Down syndrome. Although both parents were informed of the test results, they did not pursue karyotyping.

Using ultrasonography, we diagnosed a 43×32 mm supratentorial cystic structure that was located posterior to the third ventricle communicating with the widened straight sinus (Figure 1). Colour flow examination revealed turbulent flow through the lesion (Figure 2). Doppler velocity waveforms showed high blood flow through the lesion that consisted of increased peak systolic velocity and decreased resistance, which resembled an AV shunt. Investigation using 3D power Doppler revealed a much more detailed appearance of the widened vascular sinus and its communications (Figure 3). Similar vascular dilatation and turbulent flow were observed in the cervical region on colour Doppler sonography. Mild ventriculomegaly of the lateral ventricles (11 mm) was noted during the initial examination.

The ventricular dilatation increased progressively until it reached 23 mm at term.

Biometric parameters were all compatible through the first trimester sonographic examination. Mild hydramnios and significant cardiomegaly were noted. Although the patient's screening test for gestational diabetes was positive, the diagnostic test was negative and the Hb_{A1C} level was within the normal range.

The cardiac structures were normal on echocardiographic examination; however, compartments such as the aorta, pulmonary artery, and both ventricles were dilated and the heart-to-thorax ratio was increased. Periodic echocardiographic examination revealed no regurgitation at the mitral or tricuspid valve. Neither ascites nor pericardial effusion, both of which are indicators of cardiac dysfunction, was detected at any time. Parents were informed of the diagnosis of Galen vein aneurysm and neonatal complications. They opted for conservative therapy until term and surgery in the neonatal period.

A female foetus weighing 3.250 g was delivered at the 39th gestational week by caesarean section. Her Apgar scores were 7 and 7. Oxygen saturation was 70%, so the patient was intubated. Physical examination revealed dilation of both carotid arteries, hypotonic posture, and weak neonatal reflexes. She was referred to paediatric cardiology. Echocardiography was performed, and marked dilatation at the carotid arteries and its branches from the level of the arcus aorta was observed. The patient's heart circulation was hyperdynamic because of an AV fistula. Cranial magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) were performed. MRI revealed hydrocephaly and a thin parenchyma. The AV fistula was detected on MRA and the presentation was more serious than expected. Embolisation could not be performed, and the patient died on the third day after birth.

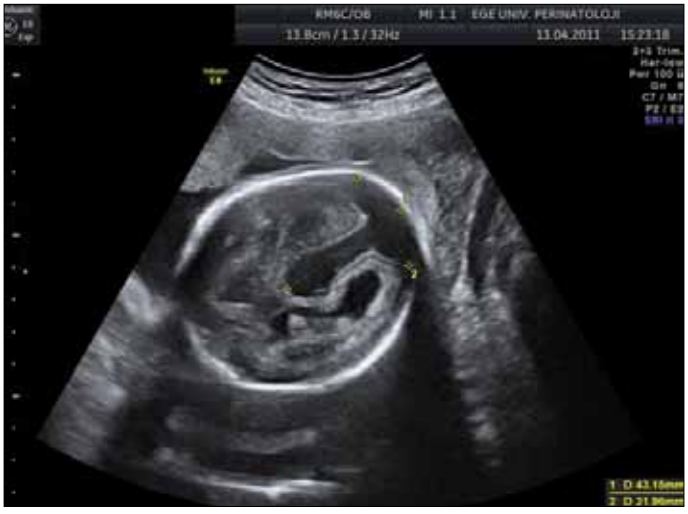

Figure 1. Cystic structure of VGA in gray scale sonography

Discussion

The current paper aimed to investigate the usefulness of 3D power Doppler sonography in a foetus diagnosed with VGAM during the second trimester. VGAM is a complex AV malformation that is usually characterized by multiple communications between the Galen vein system and the cerebral arteries (choroidal branches of posterior cerebral artery and the transmes-

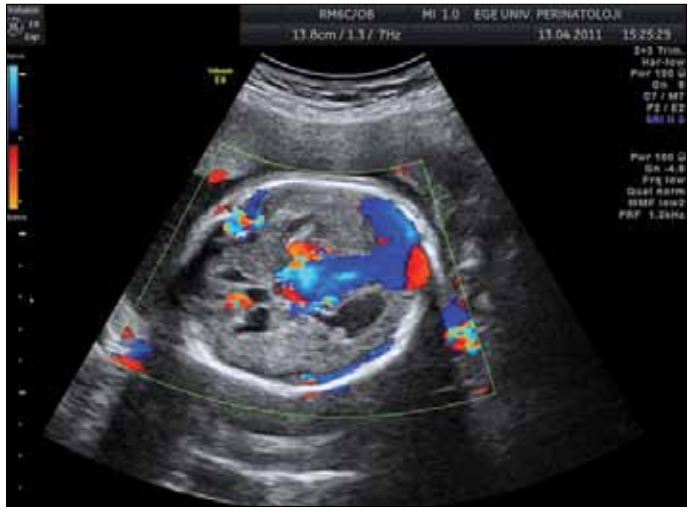

Figure 2. Turbulant blood flow in VGA through color Doppler mode

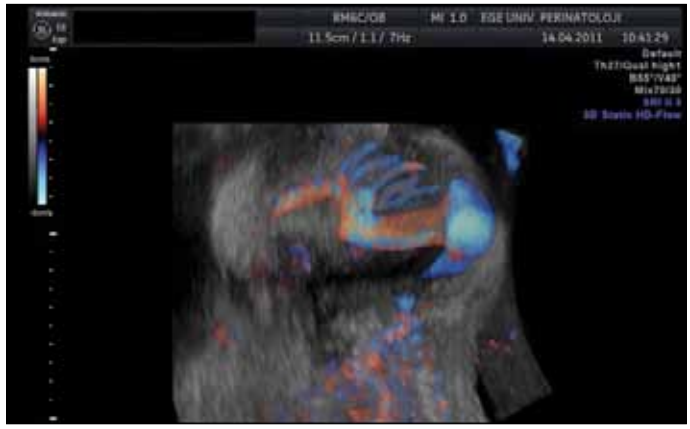

Figure 3. 3D power Doppler investigation reveals more detailed anatomic plans without disadvantages of other sonographic modes

encephalic arteries) (7). This vascular malformation may cause multi-system disturbances via hyperdynamic vascular flow or a mass effect on the intracranial structures. High output cardiac failure is the endpoint of cardiac influence. Cardiomegaly may be the initiator, and non-immune hydrops may lead to cardiac failure. In our case, the foetus was followed to term by a paediatric cardiologist to diagnose presumptive cardiac dysfunction, but no sign of cardiac pathology other than the cardiomegaly was observed.

The intracranial effects of this vascular lesion are generally directed by the mass effect of the aneurysm to the adjacent tissues such as the ventricular system. This may cause progressive dilatation of the ventricle, which was eventually diagnosed as hydrocephaly. Both hydrocephaly and diversion of the vascular flow from the cerebral cortex to the aneurysm (steal phenomenon) may cause porencephaly and leukomalacia (6). In our case, the lateral ventricle diameter increased from 11 mm to 23 mm from the initial examination to term.

In VGAM, the jugular veins may be tortuous and distended, as seen in our case. Sepulveda reported that ventriculomegaly, cardiomegaly, and jugular vein dilations were the most common findings of this pathology (10).

The diagnosis of VGAM relies on imaging studies such as ultrasonography, angiography, computed tomography, and MRI. Ultrasonography is the most common imaging method of intrauterine diagnosis. Since grey-scale sonography can only detect cystic dilatations and other peripheral manifestations, colour Doppler sonography may help to further diagnose these cystic malformations as vascular lesions. Finally, 3D power Doppler sonography shows both the nature and the anatomic structure of the lesion from different perspectives during or after examination (without the disadvantages of colour mode sonography and MRI) (11). The use of MRI during the prenatal or postnatal periods reveals detailed lesion conformation (11). Similarly, the use of angiography in the postnatal period may be helpful with diagnosis and treatment prediction.

Cardiac failure and associated brain pathologies are indicators of poor prognosis. Dilated drainage tract, multiple feeding vessels, dilated jugular veins, retrograde aortic diastolic flow, and cardiomegaly are the other factors that may influence prognosis. Hydropic changes and anatomical changes in the brain of the foetus are the worst prognostic findings. Recent studies have aimed to predict prognosis using scoring systems (12). In our case, some of the cranial reflections of poor prognosis were determined but cardiac status was stable throughout the gestational period. However, stable cardiac status does not guarantee postnatal survey. In the prenatal period, the low vascular resistance of the placenta conserves the formation of congestive heart failure by competing with the low-resistance aneurysm, but this compensated state does not predict a benign neonatal course. Neonatal congestive heart failure may exist after delivery when the placenta is removed (13, 14). The current case was similar. Although cardiomegaly was the single sign of cardiac pathology, the patient exhibited signs of cardiac failure and required intubation just after delivery.

Although treatment options have not yet been established, neurosurgical alternatives such as excision, ligation, and embolisation may be applied. Cardiac decompensation and gross brain involvement are the main indications of emergent treatment. In stable cases, treatment options are determined using a multidisciplinary approach (at 4–5 months of age). On the other hand, some cases of spontaneous regression via thrombosis have also been reported (15). In our case, both cardiac and cranial lesions were determined to have caused the mortality. In conclusion, 3D power Doppler is a non-invasive way to provide a diffuse and detailed prenatal assessment of VGAM. Although we have discovered many unknown features of VGAM, knowledge of other possible mechanisms to predict the survival of the patients with this condition is lacking.

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